A Dissertation on

A CLINICAL STUDY OF DEVELOPMENTAL GLAUCOMA

Submitted to

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With partial fulfillment of the regulations for the award of the degree of

M.S. (OPHTHALMOLOGY)

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DECLARATION

I, **Dr.B.Meenakshi,** solemnly declare that the dissertation titled **"A CLINICAL STUDY ON DEVELOPMENTAL GLAUCOMA"** has been prepared by me. This is submitted to The Tamil Nadu Dr. M.G.R. Medical University, Chennai, in partial fulfillment of the requirement for the award of M.S. Ophthalmology, degree Examination to be held in March 2008.

Place: Chennai

Date:

CERTIFICATE

This is to certify that **Dr.B.Meenakshi,** Post Graduate student in M.S Ophthalmology, at Regional Institute of Ophthalmology and Government Ophthalmic hospital attached to Madras Medical College, Chennai, carried out this dissertation on **"A CLINICAL STUDY ON DEVELOPMENTAL GLAUCOMA"** under our direct guidance and supervision during the period from May 2005 to March 2008.

This dissertation is submitted to the TamilNadu Dr.MGR Medical University, Chennai for the fulfillment of award of M.S. Degree in Ophthalmology.

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CONTENTS

INTRODUCTION

Terminologies used to represent raised intraocular pressure in children.

Developmental glaucoma

It refers to glaucoma associated with developmental anomalies of eye present at birth. It includes congenital glaucoma and glaucoma associated with other developmental anomalies, either systemic or ocular. In all forms it occurs in about 1 in 10,000 live births.

Isolated congenital glaucoma

It refers to a special form of glaucoma. These eyes have an isolated maldevelopment of trabecular meshwork not associated with other developmental ocular anomalies or ocular disease that can raise intra ocular pressure. Isolated congenital glaucoma is the most common glaucoma of infancy, occurring in 1 in 30,000 live births.

Buphthalmos or Hydrophthalmia

Buphthalmos is derived from a Greek word “ox eye” and refers to marked enlargement that can occur as a result of any type of glaucoma present in infancy. Hydrophthalmia refers to the high fluid content present with marked enlargement of an eye, which can occur in any type of glaucoma during infancy.

THEORIES OF ABNORMAL DEVELOPMENT

Von Muralt	1869	Described congenital glaucoma as one belonging to the glaucoma
Collins	1896	Abnormal mesodermal tissue in the angle
Mann	1928	Incomplete atrophy of anterior chamber mesoderm
Barkan	1955	Incomplete resorption of mesodermal cells leading to membrane formation
Allen	1955	Incomplete cleavage of mesoderm
Maumenee	1959	Abnormal anterior insertion of ciliary muscle into trabecular meshwork.
Anderson	1981	Histopathological proof that the iris and ciliary body have appearance of eye in 7th month of gestation rather than one which is at full term development.

HISTORICAL REVIEW OF SURGICAL PROCEDURES

Carlo De Vincentis	**1893**	Performed Goniotomy without visualizing angle structures.
Otto Barkan	**1938**	Perfected Goniotomy using specially designed glass contact lens to visualize angle structures.
Burian and Smith	**1960**	Performed Trabeculotomy ab externa.

NORMAL DEVELOPMENT OF THE ANTERIOR CHAMBER ANGLE

Anterior surface of the iris meets the corneal endothelium at five months of gestation to form the peripheral aspect of anterior chamber.

Slightly posterior to this junction are cells forming the developing trabecular meshwork. The ciliary muscle and ciliary process overlap the trabecular meshwork, being separated by loose connective tissue.

The trabecular meshwork later becomes exposed to anterior chamber as the angle recess deepens and moves posteriorly. The iris insertion into the angle wall is rather flat, as the angle recess has not yet formed. Posterior sliding of the uveal tissue continues during the first 6 to 12 months of life which is apparent gonioscopically as formation of the angle recess. The adult angle configuration, in which the iris turns slightly posteriorly before inserting into the ciliary body, is not normally present at birth but develops in the first 6 to 12 months of life.

EMBRYOLOGY

16 – 18 weeks: Development of scleral spur begins

Mesenchymal cells of trabecular meshwork secrete collagen and elastic tissue.

Juxtacanalicular region differentiates.

Vacuoles configuration present in endothelial lining of Schlemm's canal.

21 – 23 weeks: Angle recess at level of schlemm's canal

Trabecular meshwork consists of outer corneoscleral portion and inner uveal portion.

Corneoscleral meshwork organized as trabecular beams and inter trabecular spaces is formed.

28 – 30 weeks: Longitudinal fibres of ciliary muscle distinct and inserts into scleral spur.

Uveal meshwork shows wide intercellular space.

35 – 39 weeks: Angle recess reaches level of scleral spur.

NORMAL ANTERIOR CHAMBER ANGLE

On gonioscopy of a normal new born eye, the insertion of the iris into the angle wall is seen posterior to the scleral spur. The anterior extension of ciliary body is seen as a band anterior to iris insertion.

ANTERIOR CHAMBER IN CONGENITAL GLAUCOMA

Histological abnormalities found on examination of trabecular meshwork includes thickening of trabecular beams, thickened cords of uveal meshwork and compression of meshwork with a resultant reduction in trabecular spaces. Appearance of a membrane was due to observation of thickened compact trabecular beams in the area of meshwork adjacent to anterior chamber.

Schlemm's canal is open in early stages of congenital glaucoma. It may be obliterated in advanced stages but this is believed to be a secondary alteration from the effect of raised intra ocular pressure on ocular tissues. A thickening of juxtacanalicular connective tissue and an amorphous material in the sub endothelial area of internal wall of Schlemm's canal has been noted.

INHERITANCE

Majority of cases are sporadic

10% autosomal recessive with variable penetrance

Infantile glaucoma gene-2p21

Polygenic inheritance

CLASSIFICATION OF DEVELOPMENTAL GLAUCOMA

SHAFFER WIESS CLASSIFICATION

1. Isolated Congenital Glaucoma (Or) Primary congenital open angle glaucoma

2. Glaucomas associated with cong anomalies

    1. Aniridia
    2. Sturge Weber syndrome
    3. Neurofibromatosis
    4. Marfans syndrome
    5. Homocystinuria
    6. Lowe's syndrome
    7. Broad thumb syndrome
    8. Microcornea
    9. Microspherophakia
    10. Persistent Hyperplastic Primary Vitreous
    11. Pierre Robin syndrome
    12. Goniodysgenesis
    13. Chromosomal abnormalities

3. Acquired glaucoma (or) secondary glaucoma

a. Retrolental fibroplasia

b. Tumors—retinoblastoma, neuroblastoma

c. Inflammation— uveitis, keratitis, rubella

d. Post traumatic

HOSKINS ANATOMIC CLASSIFICATION

1. Isolated trabeculodysgenesis- Maldevelopment of trabecular meshwork.

    a. Flat iris insertion

        1. anterior insertion

        2. posterior insertion

        3. mixed insertion

    b. Concave iris insertion

2. Iridotrabeculodysgenesis

    a. Anterior stromal defects-hyperplasia/hypoplasia

    b. Anomalous iris vessels-persistent tunica vasculosa, anomalous superior vessels.

    c. Structural anomalies-holes, coloboma, aniridia

3. Corneotrabeculodysgenesis

    a. Peripheral

    b. Midperipheral

c. Central

d. Microcornea / megalocornea

GLAUCOMA ASSOCIATED WITH CONGENITAL OCULAR ABNORMALITIES

Axenfeld-Rieger syndrome

Hypoplasia/hyperplasia of iris

Peter's anomaly

Congenital ectropion uveae

Congenital corneal staphyloma

Cornea plana

Iridoschisis

Megalocornea

Morning glory syndrome

Nanophthalmos

GLAUCOMAS ASSOCIATED WITH SYSTEMIC CONGENITAL ABNORMALITIES

Weil-Marchesani syndrome

Mucopolysaccharidoses

Hallermann-Streiff syndrome

Cerebrohepatorenal syndrome (Zellweger syndrome)

Prader-Willi syndrome

Cystinosis

Oculodentodigital dysplasia

Fetal alcohol syndrome

Waardenburg syndrome

Cockayne syndrome

Stickler syndrome

RACE

No racial predilection exists.

No sex predilection exists in aniridia, Axenfeld-Rieger syndrome, Peter's anomaly, or phakomatoses.

Lowe syndrome, one of the causes of secondary congenital glaucoma, has X-linked transmission and appears in males

PATHOPHYSIOLOGY

The embryologic basis of all developmental glaucoma is fetal maldevelopment of the iridocorneal angle, called **goniodysgenesis**

TRABECULODYSGENESIS

Maldevelopment of the trabecular meshwork

IRIDOTRABECULODYSGENESIS

It is the maldevelopment of the iris anterior stroma, the iris vessels or the full iris thickness.

CORNEOTRABECULODYSGENESIS

It is the maldevelopment of the cornea which include peripheral, mid peripheral and central defects as well as microcornea and megalocornea. Peripheral corneal lesions occur adjacent to and concentric with the limbus and extend no more than 2 mm into clear cornea. These usually involve entire corneal circumference and are most often seen as posterior embryotoxon with adherent iris tissue (Axenfeld's anomaly). Mid peripheral lesions are generally found with Rieger's anomaly.

These may occur singly or in combination. Isolated Trabeculodysgenesis is the hallmark of primary developmental

glaucoma. Barkan assumed that a thin membrane covered the anterior chamber angle preventing aqueous outflow. However electron microscopic studies provided no evidence of membrane in any of the specimens. Based on clinical and histopathological observations the mechanism of developmental glaucoma has been attributed to a developmental arrest, late in gestation, of certain anterior segment structures derived from neural crest cells (Angularneurocristopathies).

High intraocular pressure causes corneal clouding, rapid enlargement of the globe and limbal stretching. The corneal diameter can enlarge up to 17mm. There may be stretching of the Descemet's membrane, resulting in linear ruptures (Haab's striae) which can lead to corneal stromal and epithelial edema as well as corneal scarring if the problem is chronic.

Sclera also expands slowly under the influence of increased IOP. The associated scleral thinning brings about visibility of the underlying uveal tissue in neonates and causes the sclera to appear blue. Thus in advanced stages the eye expands in all dimensions resembling an Ox eye (Buphthalmos).

The optic nerve head in children is more vulnerable to increased intra ocular pressure, and in advanced stages, the disc may show complete

cupping. However, optic disc cupping may be reversible with normalization of intra ocular pressure particularly in the early stage.

CLINICAL PRESENTATION

The classic **triad** of manifestation includes:

Epiphora

Photophobia

Blepharospasm

These symptoms are due to corneal irritation that accompanies corneal epithelial edema caused by elevated intra ocular pressure. Baby keeps eyes closed when exposed to light. In severe cases, the child may become irritable to the point of burying his or her head in a pillow to avoid the pain of photophobia Clouding of the cornea with or without enlargement of globe occurs. Major enlargement occurs at corneoscleral junction. As the axial length increases, myopia and astigmatism occur. Pain is unusual in older children with primary developmental glaucoma.

EXAMINATION

DIAGNOSTIC EXAMINATION

Examination under general anaesthesia (EUA) is advisable for all children who do not cooperate in an office examination.

Equipments required to perform a basic EUA

1) Pediatric speculum
2) Balanced salt solution
3) Tonometer (hand held Perkins)
4) Direct ophthalmoscope
5) Retinoscope
6) Koeppe Goniolens & light source
7) Calipers
8) Slit lamp
9) Ultrasound
10) Fundus camera

EXTERNAL EXAMINATION

The most common cause of Epiphora in the new born is blocked nasolacrimal duct (CNLDO). It is differentiated in that mucopurulent discharge is present in CNLDO.

CORNEAL ASSESSMENT

The cornea is examined to document the presence or absence of breaks in Descemet's membrane (Haab's **striae)** and corneal enlargement in order to distinguish the glaucomatous signs from other corneal abnormalities. Other causes of hazy cornea must be ruled out. These include

1) Sclero cornea
2) Tears in Descemet's membrane (obstetrical trauma)
3) Corneal ulcers
4) Metabolic diseases: Mucopolysaccharidoses & mucolipidosis
5) Peter's anomaly
6) Corneal dystrophies
7) Central corneal dermoid

CORNEAL ENLARGEMENT

The corneal diameter is measured with calipers from white to white along the horizontal and vertical meridian.

Normal neonatal horizontal corneal diameter is 10-10.5mm and it increases by 0.5 – 1mm in 1st year. Enlargement of cornea of more than **12mm** in the 1st year of life indicates glaucoma.

Conditions associated with corneal enlargement

1) Axial myopia
2) Megalocornea
3) Anterior megalophthalmos.

REFRACTION

Determination of refractive errors is done with streak retinoscopy if media is clear.

While myopia is the commonest refractive error in congenital glaucoma, its magnitude does not reach the expected value because the final refraction is influenced by

1) More flattened cornea (eyeball growth & corneal growth cause it's flattening)

2) Flattening of lens (scleral ring adjacent to the ciliary body increases in diameter – stretching of zonules – decrease in lens thickness)

3) Deep anterior chamber due to backward positioning of lens.

All these factors contribute to **emmetropisation**.

TONOMETRY

Intra ocular pressure (IOP) should be measured with a Perkins hand held applanation tonometer. In case of scarred and edematous cornea Mackay Marg tonometer is considered the most accurate.

All anesthetics agents alter IOP in the plane of anesthesia and as a direct effect on the cardiovascular tonus. A rapid lowering of IOP occurs with Halothane and readings 15-20 mm below the true measurements can be obtained.

Role of Ketamine: Recent studies show that ketamine given after premedication with diazepam and Meperidine does not raise IOP and intramuscular Ketamine even lowers IOP in children.

Standardization of anesthesia for IOP measurement for diagnosis and follow up is desirable and inconsistent reading should always be

interpreted considering the patient's general stage of anesthesia and the specific anesthetic used.

Normal IOP in an infant under Halothane anesthesia is 9-10mm Hg. IOP more than 20mm Hg indicates glaucoma. The most reliable method of measuring IOP is probably with child awake and Perkins tonometer has been found to be particularly suitable.

SLIT LAMP EXAMINATION OR EXAMINATION UNDER MICROSCOPE

This is done with a portable hand held slit lamp or binocular operating microscope.

Descemet's breaks are seen in the cornea.

Anterior chamber is characteristically deep

Iris is usually normal although it may have stromal hypoplasia with loss of crypts.

GONIOSCOPY

The Koeppe lens provides the surgeon with the appropriate view of the angle. If corneal clouding is marked, it could preclude a view of the angle. In the normal newborn eye, iris inserts posterior to the scleral spur. Trabecular meshwork appears more translucent than that of the adult. In primary congenital glaucoma the angle is usually open angle with high insertion of iris root. Iris is inserted anterior to scleral spur. The surface of trabecular meshwork may have a stippled appearance and the meshwork may be thicker than normal.

Loops of vessels from major arterial circle may be seen above iris root – **"Loch Ness monster phenomenon"**.

Peripheral iris covered by fine, fluffy tissue- **"Lister's morning mist".**

OPHTHALMOSCOPY

Cup Disc (CD) ratio greater than 0.3 or asymmetry are suggestive of developmental glaucoma. The infant glaucomatous cup is more commonly round, steep walled and central and the cup tends to enlarge circumferentially with the progression of glaucoma.

Other optic nerve head abnormalities to be ruled out are

1) Congenital malformations of the disc

2) Coloboma

3) Optic disc pit

4) Hypoplasia

5) Tilted disc of axial myopia

6) Large physiologic cup

ULTRASONOGRAPHY

Normal axial length in an infant is 17.5-20mm and increases to 22mm by 1 year. Axial length may decrease up to 0.8mm following surgical reduction of IOP.

FACTORS INFLUENCING THERAPEUTIC DECISIONS

The choice of therapy in developmental glaucoma depends on a variety of factors. The most important of these is the structural defect associated with the elevated IOP. In addition, age, corneal clarity and associated systemic syndromes can influence the choice of therapy.

A) STRUCTURAL DEFECTS

Isolated trabeculodysgenesis

It is highly responsive to goniotomy and trabeculotomy ab externa.

Irido-Trabeculodysgenesis

When other defects are associated with trabeculodysgenesis the success rate of monotony and / or Trabeculotomy is lowered. In iridodysgenesis, where the only iris defect is hypoplasia of the anterior stroma, good response to surgery has been reported.

However, when the iris defect is abnormal, vessels appear to wander irregularly across the surface of the iris, and the prognosis is extremely grave. In such cases, multiple surgeries are usually needed.

Iridocorneal dysgenesis

In patients with Axenfeld – Rieger's anomaly, surgical therapy does not have good prognosis and medical therapy is used initially. Often medical therapy too is unsuccessful, therefore, surgical intervention become necessary. In such cases, surgery should be tailored to the specific cause.

B) AGE

In general, children under the age of 3 years are best treated surgically. Children over 3 years of age deserve a trial of medical therapy unless specific defect of trabeculodysgenesis is seen.

C) CORNEAL CLARITY

In situations, where corneal clouding prevents adequate visualization of the trabecular meshwork, goniotomy is impossible. Trabeculotomy ab externa has to be performed as the initial procedure.

D) CORNEAL DIAMETER

Eyes with corneal diameter greater than 15 mm are not suitable for goniotomy. Trabeculotomy with trabeculectomy should be performed as the initial procedure.

E) **SEVERITY OF GLAUCOMA**

In advanced cases of developmental glaucoma, ab externo combined trabeculotomy with trabeculectomy should be done as it offers the highest success rate in such a situation.

MEDICAL THERAPY

Congenital glaucoma is essentially a surgical disease. Medical therapy has a supportive role to reduce the IOP temporarily, to clear the cornea and to facilitate surgical intervention.

CARBONIC ANHYDRASE INHIBITOR

Acetazolamide either alone or in combination with miotics, in a oral dose of 10-15mgs of body weight every 6 hours is safe and well tolerated by infants. It lowers IOP and reduces corneal edema as preclude to surgery. Side effects are rare.

BETA BLOCKERS

Timolol is the most commonly used medication for treatment of developmental glaucoma. It is a non selective β1 and β2 adrenergic blocker that reduces IOP by reducing aqueous inflow. Selective β blockers like betaxolol may be safer than timolol in children with asthma.

MIOTICS

Pilocarpine may be used at 1 to 2% concentration topically every 6 to 8 hours but topical application of miotics is not very effective because of the abnormal insertion of the ciliary muscle into trabecular meshwork.

SURGICAL THERAPY

GONIOTOMY

After the introduction of clinical Gonioscopy, Otto Barkan in1938 modified the Carlo Devincentis operation by using a specially designed glass contact lens to visualize the angle structures while using a knife to create an internal cleft in the trabecular tissue.

The objective of goniotomy is to incise the obstructing tissue that causes the retention of aqueous and thereby restore the access of aqueous to Schlemm's canal, thus maintaining the physiological direction of the flow. Goniotomy is most successful in the patients in whom glaucoma is recognized early and treated between 1 month and 1 year of age.

COMPLICATIONS

Hyphaema,

Iridodialysis

Cyclodialysis

Peripheral anterior synechiae

Damage to the lens

RD in high myopic eyes

TRABECULOTOMY AB EXTERNO

It is an alternative to goniotomy. It can be used even when corneal haze prevents an adequate gonioscopic view which is a prerequisite for performing goniotomy. This involves cannulating Schlemm's canal with a nylon suture at one site and threading the suture circumferentially, then withdrawing it at another site and pulling it tight like a bow string.

TRABECULECTOMY WITH TRABECULOTOMY

The trabeculotomy is performed to remove the possible obstruction to the aqueous outflow while the trabeculectomy is included to bypass the episcleral venous system. It is the procedure of choice when exact mechanism of glaucoma is uncertain. Mitomycin C increases success rate of trabeculectomy when used with caution

COMPLICATIONS

Bleb Failure

Hyphaema,

Tears in Descemets membrane,

False passage in to anterior chamber or supra choroidal space

Iridodialysis

Cyclodialysis.

Peripheral anterior synechiae.

Injury to lens

Staphyloma

AQUEOUS SHUNTING PROCEDURES

Device is made of non-reactive synthetic material to which fibroblast adheres poorly. The bleb is placed away from the anterior limbus to reduce inflammation and thinning.

Pediatric Molteno implants (8mm in diameter) and baby Baerveldt implants are specifically designed for this purpose.

COMPLICATIONS

Hypotony

Tube endothelial touch

Erosion of tube

Migration of tube

Cataract formation

CYCLODESTRUCTIVE PROCEDURES

If the IOP is unduly high, rate of aqueous production is reduced by injuring the ciliary processes preferably by Cyclocryotherapy, or Nd-YAG laser transscleral photocoagulation.

TREATMENT OF COMPLICATIONS

Persistent corneal clouding after reduction of IOP is treated by PKP Occlusion therapy is done in case of amblyopia.

LONG TERM FOLLOW UP

Between 3 to 6 weeks after surgery the post operative control of the glaucoma must be judged. The degree of relief from photophobia, tearing and blepharospasm usually reflect the effectiveness of surgery and may reasonably predict whether additional surgery is required.

Each follow up evaluation include :

Visual acuity testing

External examination

Appearance of the filtering bleb

Corneal assessment

Ophthalmoscopy

Refraction

Gonioscopy

Tonometry

Ultrasound biometry.

VISUAL REHABILITATION

Visual rehabilitation is as important in the management of the disease as is the control of IOP. Visual rehabilitation involves correction of refractive errors, correction of opacities in the media such as corneal scarring and cataract, and orthoptic treatment to stimulate the development of binocular stereoscopic vision. Anisometropia and amblyopia must be aggressively treated to give good vision in both eyes. These should be undertaken as early as possible.

AIM OF THE STUDY

The aim of the study is to analyze

1) Average age of presentation
2) Sex distribution
3) Association with consanguinity
4) Laterality
5) Associated systemic and ocular anomalies
6) Complications
7) Mode of treatment and outcome of developmental glaucoma

INCLUSION CRITERIA

Developmental glaucoma in the pediatric age group (from birth to 12 years of age)

EXCLUSION CRITERIA

1) Traumatic glaucoma
2) Post inflammatory glaucoma
3) Other causes of epiphora
4) Other causes of corneal haziness without raise in intra ocular pressure
5) Other causes of megalophthalmos with out rise in intra ocular pressure

MATERIALS AND METHODS

This is a prospective study conducted in Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Chennai during the period of June 2005 to September 2007.

66 eyes of 37 patients were included in the study. Children who were not cooperative for office examination were examined under ketamine anesthesia.

Parameters that were recorded included:

1) Age of presentation
2) Sex of the individual
3) History of consanguinity
4) Eye involved
5) Association with any systemic and /or ocular anomalies
6) Vision
7) Under ketamine anesthesia intra ocular pressure was recorded using Perkins tonometer. Corneal diameter was measured using Vernier calipers. Fundus examination was done using direct ophthalmoscopy
8) Axial length

9) B Scan was done in cases were there was no view of the fundus.

After diagnosing developmental glaucoma, patients who were high risk for surgery under general anesthesia were started with medical management.

Surgeries were done once patients obtained GA fitness. All the surgeries were done by a single experienced surgeon.

25 eyes were started with medical therapy. 40 eyes underwent surgery, of which 21 eyes underwent Trabeculectomy, 15 had trabeculotomy with trabeculectomy, 1 eye had trabeculectomy with deroofing of schlemm's canal, and 1 eye associated with Weil Marchesani syndrome underwent Extra capsular cataract extraction (ECCE) with Anterior vitrectomy.

Post operatively corneal clarity was assessed. Repeat intra ocular measurements were done on follow up.

Patients with failure of surgery underwent repeat surgery.

Repeat surgeries included trabeculotomy with trabeculectomy, trabeculotomy with trabeculectomy with Mitomycin C. Patients with failure of second surgery underwent third surgery. Patients were followed up and refractive errors were corrected and spectacles were prescribed if possible. Cases with amblyopia were started on occlusion therapy.

OBSERVATION AND DISCUSSION

AGE DISTRIBUTION

Table 1

Age	<1 yr	13-36 months	37-60 months	5-7 yrs
Number	14	12	7	4
percentage	38	32	19	11

The most common age of presentation in this study was below 1 year of age followed by 1-3 years of age. This is comparable with the study conducted, at Department of ophthalmology, Royal Children's Hospital and Centre for Eye Research Australia, Department of Ophthalmology, University of Melbourne, Royal Victorian Eye and Ear Hospital, Melbourne. The mean +/- SD age at presentation in this study was 135 +/- 84 days.

SEX DISTRIBUTION

Table 2

Sex	Male	Female
Number	13	24
Percentage	35	65

In this study female children were more commonly affected which constituted about 65% of cases

CONSANGUINITY

Table 3

Consanguinity	Non consanguinous	1st degree	2nd degree	3rd degree
Number	32	nil	3	2
Percentage	86	nil	8	6

Most of the children in this study were born of nonconsanguinous marriage.

LATERALITY

Table 4

Laterality	BE	RE	LE
Number	29	6	2
Percentage	78	16	6

Most of the cases had bilateral involvement. Cases which had unilateral involvement were mostly due to phakomatoses.

ASSOCIATED SYSTEMIC ANOMALIES

Table 5

Systemic anomalies	Number
Sturge Weber	4
Mucopolysaccharidoses	2
Congenital heart disease	3
Microcephaly with seizure disorder	2
Oculomelanocytosis of OTA	1
Cleft lip with cleft palate	1
Down syndrome	1

14 cases in this study had associated systemic anomalies. Of this 4 cases had Sturge Weber syndrome

ASSOCIATED OCULAR ANOMALIES

Table 6

Ocular anomalies	Number
Weil marchesani	1
Reiger's anomaly	1

2 cases had associated ocular anomalies of which 1 had Weil Marchesani and the other had Rieger's anomaly

TENSION

Table 7

Tension recorded (mm of Hg)	Number of eyes	Percentage
<19	3	5
20 – 30	43	65
31 – 40	9	14
41 – 50	10	15
>51	1	1

Of the 66 eyes that were studied 43 eyes had tension recording between 20 and 30 mm of Hg. Only 1 eye had tension more than 51 mm of Hg. Tension below 20 mm of Hg was recorded in cases with Retinal Detachment (RD). One case with RD had associated choroidal detachment and had gone for phthisis bulbi. The second case of

RD had corneal diameter of 16mm and an axial length of 30 mm. This can be compared with a study paper presented at the annual meeting of American Academy of ophthalmology in November 2001 in which the mean pre operative IOP was 26.9+/- 5.2 mm of Hg (range – 14 – 42).

AXIAL LENGTH

Table 8

Axial length	Number of eyes	Percentage
<21.9	3	5
22 – 23.9	49	74
>24	14	21

Majority of cases had axial length of 22 – 24 mm. 14 cases had axial length of more than 24 of which the highest recorded was 30 mm and this case had an associated Retinal detachment. This is comparable with the study conducted at University Eye Hospital Wuerzburg, Joseph – Schneider, Germany published in September 2006. In this study the mean axial length of the eye ball at first presentation was 22.6+/- 1.8mm.

REFRACTION

Table 9

Refractive Error	Number of eyes	Percentage
Myopia	32	47
Hypermetropia	7	11
Emmetropia	9	16
No view	17	26

Most common refractive error was myopia which was present in 32 eyes, followed by emmetropia in 9 eyes. Hypermetropia occurred in 7 eyes.

This is comparable with the study paper presented at the annual meeting of the American Academy of ophthalmology. In this study myopia was the most common refractive error presenting in approximately half of the eyes.

In another paper presented at the association for research in vision and ophthalmology meeting at Florida, myopia was the most common refractive error accounting for 75% of cases.

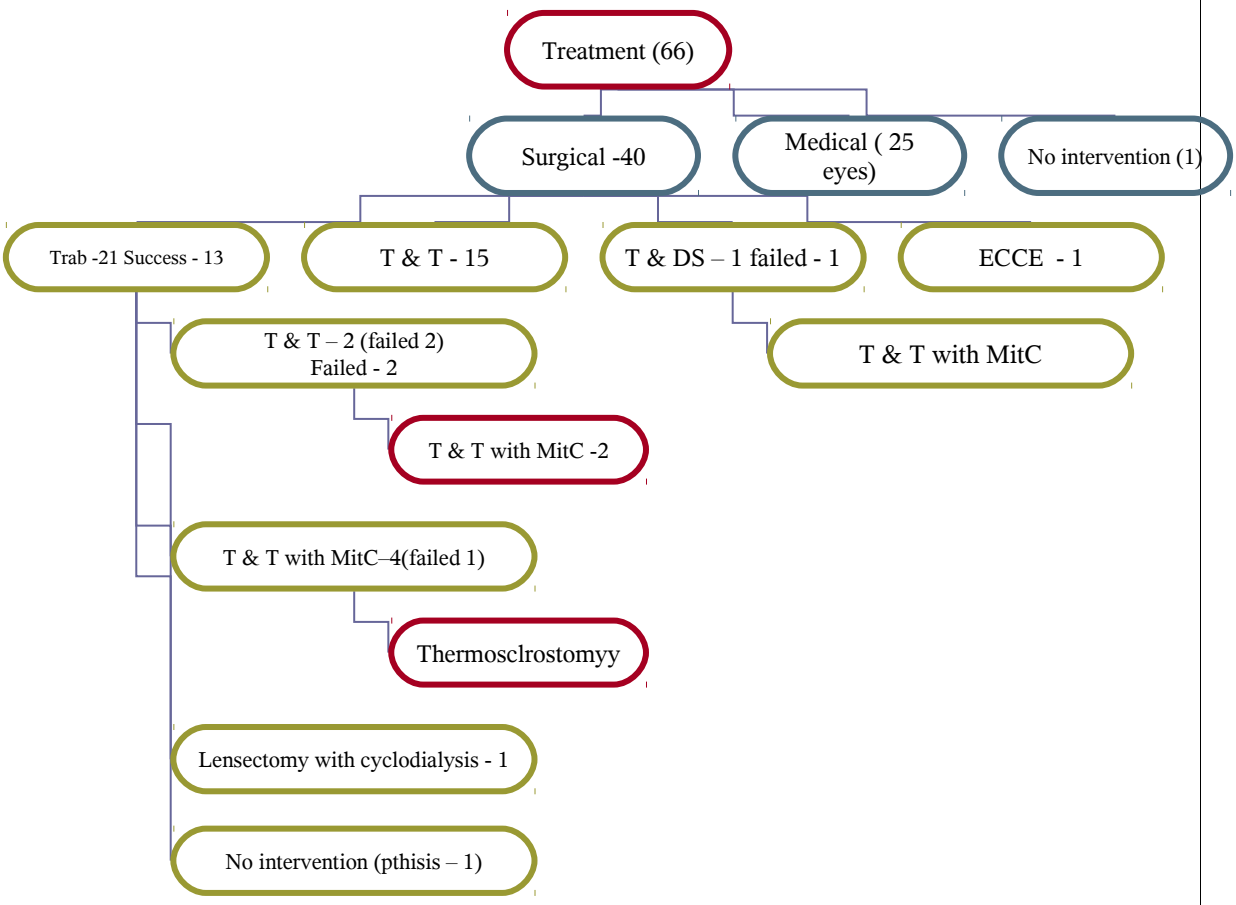
COMPLICATIONS

Complications of developmental glaucoma that were encountered in this study were as follows

Table 10

Complications	Number of eyes
Corneal decompensation	17
Cataract	4
Retinal detachment	1
RD with choroidal detachment	1
Phthisis	2

TREATMENT


Treatment (66)
Surgical -40
Medical (25 eyes)
No intervention (1)
Trab -21 Success - 13
T & T - 15
T & DS – 1 failed - 1
ECCE - 1
T & T – 2 (failed 2) Failed - 2
T & T with MitC
T & T with MitC -2
T & T with MitC–4(failed 1)
Thermosclrostomyy
Lensectomy with cyclodialysis - 1
No intervention (pthisis – 1)

MEDICAL TREATMENT

Of the 37 patients, 13 (25 eyes) received medical treatment. Of these, 2 patients were not willing for surgery. 1 patient had cleft lip with cleft palate. 2 patients had seizure disorder. 3 patients had congenital heart disease. 4 patients were less than 3 months of age. 1 eye was phthisical.

SURGICAL TREATMENT

40 eyes had surgical treatment. Of these trabeculectomy was done as primary procedure in 21 eyes. Of the 21 eyes, 13 eyes had successful control of intra ocular pressure and 8 failed. Of the 8 eyes, 4 eyes underwent trabeculotomy with trabeculectomy with Mitomycin C, 2 eyes were taken up for trabeculotomy with trabeculectomy, 1 case underwent lensectomy with cyclodialysis. 1 eye became phthisical.

Of the 4 eyes which underwent Trabeculectomy with Trabeculotomy with Mitomycin C, 3 were successful and 1 case which had oculomelanocytosis of OTA underwent a third surgical procedure - Thermosclerostomy.

Of the 2 eyes which had undergone trabeculotomy with trabeculectomy, both failed. Repeat trabeculotomy with trabeculectomy with Mitomycin C was done.

Trabeculotomy with trabeculectomy was done as primary procedure in 15 eyes. All cases which underwent Trabeculectomy with trabeculotomy as the first procedure were successful until the follow up period.

1 eye had trabeculectomy with deroofing of Schlemm's canal as the primary procedure and this failed. Repeat trabeculotomy with trabeculectomy with Mitomycin C was done for this case.

A case of Weil Marchesani syndrome underwent Extra Capsular Cataract extraction with anterior vitrectomy as the primary procedure and this procedure was successful in this case.

2 cases with painful blind eye were enucleated.

Post operative clearing of cornea and reduction of intra ocular pressure were taken as parameters to decide whether the procedure was successful.

Table 11

Treatment Modality	Number of eyes
Medical	25
Surgical	40
Phthisical without treatment	1

Table 12

TYPE OF SURGERY DONE AND SUCCESS RATE:

PRIMARY PROCEDURE

Type of surgery	Number of eyes	Success	Failure
Trabeculectomy	21	13	8
Trabeculotomy with Trabeculectomy	15	15	
Trabeculectomy with deroofing of Schlemm's canal	1		1
ECCE with Anterior vitrectomy	1	1	
Enucleation	2		

Repeat surgery was done in the failed cases.

Table 13

Second surgery

Type of surgery	Number of eyes	Success	Failure
Trabeculotomy with trabeculectomy with Mitomycin C	4	3	1
Trabeculotomy with trabculectomy	2		2
Lensectomy with cyclodialysis	1	1	
No intervention (Phthisis bulbi)	1		

A third surgery was done for 3 eyes

Table 14

THIRD SURGERY

Type of surgery	Number of eyes	Success	Failure
Thermosclerostomy	1	1	-
Trabeculotomy with trabeculectomy with Mitomycin C	2	2	-

VISUAL OUTCOME

17 eyes of 11 patients had visual acuity greater than 6/60, of which 5 eyes had visual acuity greater or equal to 6/12 with correction.

5 eyes of 4 children had vision of No PL and 2 eyes had been enucleated.

12 infants (22 eyes) fixed and followed light. Vision recording was not possible in 3 infants (6 eyes) as they were less than 1 month of age.

14 eyes of 7 patients had vision of less than 6/60 of which 3 children had only PL. of the 3 children who had PL, 2 had defective PR.

Table 15

Vision	Number of patients
No PL	5
Fixing & Following light	22
PL+ to 6/60	14
6/60 – 6/12	12
6/12 – 6/6	5

In the study paper presented in the American Academy of Ophthalmology 26.3% had final best corrected visual acuity of 6/12, 36.8% had more than 6/ 18 and 42.1% had final visual acuity less than 6/18 to 6/60.

FOLLOW UP

Cases were followed up for 3 months, 6 months, 1 year depending on the time of surgery.

Table 16

Type of surgery	Follow Up			
		3 months	6 months	1 year
Trabeculectomy	Number of eyes	21	17	13
	Success	17	13	13
	Failure	4	3	-
Trabeculotomy with Trabeculectomy	Number of eyes	17	15	15
	Success	16	11	9
	Failure	1	1	
Trabeculotomy with Trabeculectomy with Mitomycin C	Number of eyes	4	4	4
	Success	4	4	3
	Failure			1
Thermosclerostomy	Number of eyes	1	1	1
	Success	1	1	
	failure			
Lensectomy with cyclodialysis	Number of eyes	1	1	1
	Success	1	1	1
	Failure			
ECCE with Anterior vitrectomy	Number of eyes	1	1	1
	Success	1	1	1
	Failure			

TRABECULECTOMY

At the end of 1 year, out of 21 eyes which underwent trabeculectomy 13 eyes had good control of intra ocular pressure. 8 eyes which underwent Trabeculectomy had failed. Of the 8 eyes, 7 were taken up for repeat surgery and 1 eye became phthisical.

TRABECULOTOMY WITH TRABECULECTOMY

At the end of 1 year of follow up 9 cases out of 17 cases which underwent trabeculotomy with trabeculectomy had successful control of intra ocular pressure. 2 cases had failed.

TRABECULOTOMY WITH TRABECULECTOMY WITH MITOMYCIN C

Out of 4 cases followed up for 1 year 3 had good control of IOP. 1 case had failed at end of 1 year.

The success rate was 81%, 67% and 67% at the end of 3 months, 6 months and 1 year respectively for trabeculectomy. The success rate for trabeculotomy with trabeculectomy at the end of 3 months was 88%. The follow up period of trabeculotomy with trabeculectomy was too short to be compared with other studies.

In this study all 4 cases of Sturge Weber syndrome which underwent Trabeculotomy with Trabeculectomy were successful and were managed without additional medical therapy. All 4 cases had good visual acuity.

In an article presented in American Academy of Ophthalmology April 2000 all cases of Sturge Weber syndrome who underwent Trabeculotomy with Trabeculectomy were successful.

In another Polish article published on efficacy of treatment in glaucoma associated with Sturge Weber syndrome, 4 patients with Sturge Weber syndrome were studied. 2 patients underwent trabeculectomy and 2 underwent non penetrating deep sclerectomy with SK Gel implant. After surgery the IOP was not under good control and 2 patients required 2 medications and 2 patients required 1 medication.

POST OPERATIVE IOP

Of the 40 eyes operated, 31 eyes had IOP less than 20 mm of Hg and 7 eyes had IOP between 21 and 25 mm of Hg. This was comparable with the study paper presented at the annual meeting of American Academy of ophthalmology in November 2001 where the mean post op IOP was 14.5+/- 3.8 mm of Hg (range 8-28).

Table 17

Post op IOP mm of Hg	Number of patients	Percentage
Upto 20	31	82
21 -25	7	18

RESULTS

Most common age group that affected was less than 3 years which accounted for 70% of cases.

Female children were more commonly affected than male children in this study. 75% of cases were female.

There was no association with consanguinity. 78% of cases had bilateral involvement.

Most common systemic anomaly was Sturge Weber syndrome seen in 4 cases, followed by congenital heart disease which accounted for 3 cases.

Most common complication associated with developmental glaucoma in this study was corneal decompensation which occurred in 17 eyes followed by cataract in 6 eyes. Retinal Detachment occurred in 2 eyes and 2 eyes became phthisical.

Myopia was the most common refractive error. 45% of cases had myopia.The success rate was 81%, 67% and 67% at the end of 3 months, 6 months and 1 year respectively for trabeculectomy. The success

rate for trabeculotomy with trabeculectomy at the end of 3 months was 88%.

21 eyes had poor visual outcome of vision less than 6/60 of which 6 eyes had No PL and 2 eyes were enucleated.

16 eyes had visual acuity better than 6/60 of which 5 eyes had visual acuity better than 6/12.

CONCLUSION

Developmental glaucoma is a global problem. A proper diagnostic evaluation under general anesthesia is required. Surgery remains the principle therapeutic modality in the management of developmental glaucoma. Trabeculotomy is valuable in the management of primary developmental glaucoma, while combined Trabeculotomy with Trabeculectomy offers the best hope of success in advanced cases. Trabeculotomy with trabeculectomy with Mitomycin C proves very useful in cases with failure of primary procedure. It is important that the appropriate operation should be chosen and performed with technical perfection in a tertiary care centre where there is adequate facility to ensure safe anaesthesia and skillful surgery with a lifetime follow up.

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PROFORMA FOR DEVELOPMENTAL GLAUCOMA

Name of the patient: IP No:
Age: Sex: Informant:
Place:

Complaints: Opacity in the black portion of the eye
Photophobia
Watering of eyes
Prominence of eyes

Antenatal history:

Consanguinity:

Birth history: FTND/ LSCS
Preterm
Birth asphyxia
Birth trauma
Congenital anomalies

Developmental history:

Siblings:

Family history:

Treatment History:

General Examination:
Congenital anomalies:
Facial asymmetry:

Systemic Examination:
Higher functions
CVS
RS
P/A

Ocular Examination: RE LE

- Blepharospasm
- Vision
- Lids
- EOM
- Conjunctiva
- Cornea
- Anterior chamber
- Iris
- Pupil
- Lens

Examination under general anaesthesia:

- Corneal diameter:
  - Horizontal
  - Vertical
- Axial Length
- Tension:
- Fundus:
  - RE
  - LE

Diagnosis:

Treatment:

- Medical:

- Surgical:

KEY TO MASTERCHART

S.NO	-	SERIAL NUMBER
RE	-	RIGHT EYE
LE	-	LEFT EYE
BE	-	BOTH EYES
COR DIA	-	CORNEAL DIAMETER
REF	-	REFRACTION
COMPLI	-	COMPLICATIONS
CL	-	CLEFT LIP
CP	-	CLEFT PALATE
SWS	-	STURGE WEBER SYNDROME
HS	-	HURLER'S SYNDROME
MC	-	MICROCEPHALY
S	-	SEIZURE DISORDER
MPS	-	MUCOPOLYSACCHARIDOSES
ASD	-	ATRIAL SEPTAL DEFECT
OTA	-	OCULODERMAL MELANOCYTOSIS OF OTA
CHD	-	CONGENITAL HEART DISEASE
DS	-	DOWNS SYNDROME
WM	-	WEIL MARCHESANI
RA	-	REIGER'S ANOMALY
NP	-	NOT POSSIBLE
HM	-	HAND MOVEMENTS
F&F	-	FIXES AND FOLLOWS LIGHT
NV	-	NO VIEW
M	-	MYOPIA
H	-	HYPERMETROPIA
E	-	EMMETROPIA
CO D	-	CORNEAL DECOMPENSATION
C	-	CATARACT
RD	-	RETINAL DETACHMENT
CH D	-	CHOROIDAL DETATCHMENT
TIM	-	TIMOLOL
B	-	BETOXALOL
T	-	TRABECULECTOMY
T & T	-	TRABECULOTOMY WITH TRABECULECTOMY
T & T & MMC	-	TRABECULOTOMY WITH TRABECULECTOMY WITH MITOMYCIN C
L & C	-	LENSECTOMY WITH CYCLODIALYSIS
T & DS	-	TRABECULECTOMY WITH DEROOFING OF SCHLEMM'S CANAL
ENU	-	ENUCLEATION